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FACIAL PARALYSIS: A STUDY OF THREE HUNDRED AND THIRTY-FIVE CASES[1]

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About five years ago, being impressed by the frequency with which patients, suffering from facial paralysis, were presenting themselves at the Out Patient Department of the Massachusetts General Hospital, I became interested in following a series of cases, in an attempt to determine whether those in which a complete R. D. was found made a more rapid or more perfect recovery when treated by galvanism. In pursuing this line of investigation, various questions suggested themselves for solution, viz.: Is exposure to cold an important factor in the etiology? Do the attacks occur with much greater frequency in the winter months? Are the patients, affected in the later years, more liable to the severe form of paralysis than the young or middle-aged? Does continued galvanic treatment play any rôle in the predisposition to secondary contraction? Is the initial pain of any prognostic value regarding the severity of the attack?

In order to study a sufficient number of cases, to make these statistics of value, I have attempted to see and examine personally all the patients coming to the neurological department for facial paralysis during the past five years and have made use of the hospital records of such cases for the three years preceding this period. To this number of cases I have added the data of those which I have seen in my practice and through the courtesy of my colleagues, making in all a series of 335. This number represents only the neuritic type of facial paralysis, all cases due to ear disease, cerebral syphilis, fracture of the base, tumors and glands being excluded.

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The question as to whether males are more frequently affected than females, has been considered by numerous investigators in studying the condition. The greater exposure necessitated by the occupations of men, would lead one to expect men to be more prone to the disease than women, and while this is literally true, the variation between the two sexes is so slight that it needs no theory as to its cause.

Gowers (1) in 112 cases found 66 in males and 46 in females; Bernhardt (2) in a series of 57 cases, published in 1888, found 34 in men and 23 in women, while in a second series, published four years later (3), he found 32 men and 23 women. Hübschman (4), in 99 cases, treated during 14 years in the University Polyclinic at Leipsig, found 54 males and 45 females. Sossinka (5), in his inaugural dissertation at Leipsig, reported 300 cases of facial paralysis from Mendel's Polyclinic at Berlin; unlike the above authors, he found a preponderance of cases among females in the ratio of 172 females to 128 males.

My own series of 335 cases shows 191 males and 144 females. Taking the combined sums of these series, it gives us a total of 968 cases, and shows that among this number of patients 52 per cent. were male and 48 per cent. female; a difference which can almost be considered negligible.

Whether the right or left side is more frequently affected, can have no especial importance other than to be of passing interest. Bernhardt (3) found 63 of his cases of paralysis to be right-sided and 49 to be on the left, while Hübschman (4) found 48 to be on the left and 47 on the right side. In those cases in which the side paralyzed is recorded in my series, the right nerve was affected 163 times and the left 157 times, while in two instances both sides were involved. There, as in the consideration of the sexes, the difference is so slight, compared with the number of cases considered, that we may conclude the two sides equally liable to paralysis.

The changes occurring in the nerve in facial paralysis have now been fairly extensively studied by a number of investigators, and the conditions found in the different cases show very little variation in the pathology. Minhowski (6) in 1890 was the first to describe the pathology of the condition, and called attention to the fact that this process was one of simple parenchymatous degeneration without swelling. This was corroborated by the report of Dejerine and Theohari (7) in 1897, and Myer (8) in the same year described the condition in this country. Since then, Mirallié (9), Alexander (10) and André Thomas (11) have all described essentially the same condition.

The degeneration of the axis cylinders is found in the peripheral ramifications of the nerve, and persists as far up as the geniculate ganglion, although the trunk shows no evidence of an inflammatory process. André Thomas (11) found the axis cylinders to be somewhat swollen beyond the geniculate ganglion, although they became rapidly normal in appearance as they approached the medulla. The cells of the facial nucleus are uniformly described as being in various stages of chromatolysis.

If then the process causing facial paralysis is a parenchymatous degeneration, such as we find in toxic forms of neuritis, rather than an inflammatory condition, what are we going to accept as the predisposing factors of the attack?

Neumann (12) has maintained that an hereditary neuropathic taint plays an important part in predisposing the individual to an attack. He gives the histories of 37 patients who suffered from Bell's paralysis and showed that in 24 of these patients there was a record of nervous disease in the near relatives. Insanity was found in ten families and epilepsy in seven. In five of his cases, facial paralysis had occurred in other members of the family: in one three sisters suffered from attacks, and in another, three sisters, together with a child and a grandchild of one, had been affected. This series of Neumann is certainly a remarkable one and in itself is significant. It has been much quoted, and Despaigne (13), in his treatise on the subject, expresses himself as also believing that a neuropathic family history predisposes to attacks. I must confess, however, that I am not in sympathy with this view, for many inquiries on this point with patients has led me to believe that neuropathic relatives are no more frequent in families of those stricken with facial paralysis than are found in families taken at random.

Williams (14) in a report of a case of facial paralysis, appearing in a woman the day after her child fell from her arms, suggests that fright may play a part in the causation of an attack. I have twice seen attacks follow emotional excitement; in one instance the attack came the day after the patient had been run away with by a horse, and in the other case it followed the shock of a sudden death in the patient's family. Such instances are exceptional, however, and may be regarded more in the nature of a coincidence than as illustrative that emotional disturbance is an etiological factor in facial paralysis.

The fact that attacks may have a tendency to recur in the same individual, or may appear in other members of the family, is explained by Sarbo (15) on the ground that there may be an inherited anomaly or narrowing in the bony canal of the nerve. He thinks the paralysis is due, not to an infection, but rather to an alteration in circulation in the nerve, giving rise to pressure.

Stenger (16), in a collection of cases, demonstrated a large aural canal, which he believes allows free access to the cold, so that the ear becomes affected and causes a secondary involvement of the facial nerve. He thinks that the early ear symptoms are frequently overlooked by the physician.

Reik (17) also considers the ear to be of importance in the etiology of the neuritic form of facial paralysis. He has made a study of 300 temporal bones and finds that one-half of these have a perforation from the tympanic cavity to the Fallopian canal, other than the two normal ones for the chorda tympani and the stapedius branch of the nerve. In this way exposure to cold causes an acute otitis media with inflammatory swelling of the mucous membrane, which may extend through the perforation or a dehiscence in the canal wall to involve the facial nerve. He examined 12 cases from Dr. Thomas' clinic, 10 of which had had initial aural pain, and found positive evidence of middle ear disease in all cases. In conclusion, he makes the claim that " In the majority, if not all, cases of refrigeratory facial paralysis, an acute or subacute otitis media is an intermediary condition between the exposure to cold and the appearance of the paresis."

Since the publication of Reik's paper four years ago, I have made a special effort to have all cases of facial paralysis, presenting themselves early in the course of the disease, referred to an aurist, for a report as to the condition of the middle ear. Of course a large majority of the cases do not seek hospital aid until after the first week, so that any ephemeral congestion of the ear may have disappeared at the time of the out-patient's examination. For this reason I have included, for the determination of aural complication, only those cases in which the examination was made during the first seven days. In twenty-two cases reports have been returned from the aurist, nineteen of which declared the middle ear to be normal, while three described a mild degree of congestion, a ratio which is far from being in accord with Reik's findings.

Whether the neuritis is a primary one, or is secondary to an inflammatory process of the middle ear, there can be no doubt that exposure to cold winds and draughts plays a part in the causation of an attack. Neumann (18) rejects this theory, however, and maintains that the factors acting to cause an attack of facial paralysis are: (1) A diminished power of resistance of the nerve, acquired or hereditary; (2) the action of substances on the nerve as urea and uric acid, etc., or the toxines of such diseases as syphilis or diabetes; (3) diminished power to act against these substances on account of the lessened resistance of the protecting cells of Schwann's sheath and the connective tissue. He believes that this resistance may be lowered by shock or trauma, or from injury, resulting from the obstruction to the flow of secretion from the glands which surround the nerve at its point of exit from the stylomastoid foramen.

The theory that the action of some toxic substances on the nerve plays an important role in the etiology of attacks, seems to be borne out by the fact that a retrobulbar optic neuritis may occur in individuals who have previously been affected with facial paralysis. Such cases have been reported by de Schweinitz (19) and Schumway (20) and, although in all three cases described, the optic neuritis did not manifest itself until one or two years after the facial paralysis, it may be taken as an indication that the patient was liable to such toxic agents.

Although it is true that paralysis of the facial muscles often occurs without a history of direct exposure to cold, I am convinced that this history is to be too often obtained to admit the possibility of coincidence. If, however, the degree of exposure or intensity of the cold is the primary cause, we should expect to find attacks of facial paralysis to occur more frequently in the winter months. Gowers (1) states that attacks are more frequent in the winter than in the summer months. Bernhardt (3) and Hübschman (4) have both given tables, illustrating the months of onset of attacks, dividing their cases into mild, medium and severe classes. These results I have tabulated below, without reference to the severity of the disease, together with my own.

	Bernhardt.	Hübschman.	Waterman
January	7	7	27
February	3	9	23
March	1	3	28
April	5	7	22
May	5	2	22
June	6	9	32
July	2	7	29
August	5	3	23
September	7	8	33
October	1	7	30
November	6	8	27
December	5	7	24

A division of my cases into the mild type, as contrasted with those showing a partial or complete reaction of degeneration, is also illustrated by the following table. Some of the cases, tabulated above, do not appear in this division however, since the hospital records occasionally fail to show a record of the electrical examination.

	Mild.	Severe.
January	6	17
February	7	15
March	14	9
April	7	12
May	5	14
June	13	17
July	9	14
August	5	13
September	10	20
October	6	18
November	2	15
December	5	16

It is apparent from these figures that the degree of cold is not the important factor in the causation of facial paralysis, for were it so, in a climate such as ours, where the extremes of temperature are so great in the summer and winter, we should expect a considerable difference in the number of cases appearing in these seasons. As a matter of fact, not only does the average of the different months seem to be almost a uniform one, but neither do the mild cases appear more frequent in the summer, nor are the severe ones more numerous in the winter months. We must conclude therefore, as Bernhardt (3) has suggested, that though exposure may precipitate an attack, the real cause lies in the lessened power of resistance of the individual.

That one attack predisposes the individual to subsequent attacks, was a theory advanced by Eulenberg (21), who cited a case of a man who had three times had facial paralysis on the left side and twice on the right. Numerous such instances of recurrent attacks in the same person are to be found in the literature on the subject, but it seems more probable that the individual is anatomically predisposed, or has a low power of resistance to the toxic substance causing the attack, than that the first attack should predispose to later ones. It is probably this same predisposition, existing in other members of a family, which gives rise to the occurrence of several attacks in the same household as in the instance described by Arkwright (22), who reported three sisters and one brother being afflicted with facial paralysis at the ages of 16, 16, 17 and 26.

It is not my intention to deal in detail with the recurrent and diplegic cases, for this aspect of the disease has been too well worked out and reported by others, notably by Bernhardt in 1899, and more recently by (Émile Paul) Petit (23). He has made an analysis of the previously reported cases of recurrent facial palsy and adds twelve of his own, observed at the Charcot Clinic at the Salpêtrière. These cases are all carefully studied regarding the severity of secondary and tertiary attacks, the periods elapsing between attacks and the prognosis of such seizures. Petit estimates that six per cent. of all cases of facial paralysis have a recurrence. In my series 2.7 per cent. suffered a recurrence. Eight of these gave a history of one previous attack and one was suffering from his third seizure when seen.

However great may be the rôle played by infection and toxic substances in the causation of facial paralysis, it seems probable that the age of the individual, at the time of the onset of the disease, directly influences the severity of the attack. The following table illustrates the age at which the severe and mild attacks have affected the cases which we are studying. Unfortunately the failure to give the electrical reactions in some of the records makes it possible to record only 282 cases in this tabulation:

	187 Severe Cases.	95 Mild Cases.
0 to 10 years......	6 per cent.	3 per cent.
10 to 20 years......	10 per cent.	20 per cent.
20 to 20 years......	31 per cent.	36 per cent.
30 to 40 years......	19 per cent.	27 per cent.
40 to 50 years......	17 per cent.	6 per cent.
50 to 60 years......	12 per cent.	4 per cent.
60 tô 70 years......	3 per cent.	2 per cent.
70 to 80 years......	2 per cent.	2 per cent.

Of course statistics of most diseases show that the percentage of those from sixty to eighty years of age affected is relatively low. But the greater per cent. of severe attacks between the ages of forty and sixty, as compared with the milder form of the disease, seems sufficiently striking to be significant. Is it not probable that the resistance of the organism to this disease, already low in those affected, is further diminished by advancing years?

The relation of the initial pain and the premonitory herpes to the facial paralysis has for many years been discussed, but no satisfactory explanation has been offered for their occurrence until 1906, when Hunt (24) read his paper before this society on "Herpetic Inflammations of the Geniculate Ganglion." The part played by the geniculate ganglion and sensory fibers of the facial nerve, described by him, must be accepted, and it is only in considering the question of prognosis that I wish to refer to the severe pain, which so often precedes or ushers in an attack of paralysis. Is the initial pain of any prognostic value in determining whether the motor paralysis is to be of a mild or severe type? Eulenburg was of the opinion that the intensity of the pain may be an indication of the severity of the paralysis to follow, a view with which Moebius (25) coincided. Testaz (27), in a thesis entitled "Paralysie douloureuse de la septième paire," expressed himself as believing that the pain was of direct prognostic value in determining the severity of the attack. Bernhardt (3), however, called attention to the fact that pain may be absent in the severe form of facial paralysis, while it may be a prominent symptom in the early stages of a light attack. Menke (26), from a study of 105 collected cases, also made this observation, while Hübschman (4) in 1894 found that in his series of cases there was a history of pain only once in the sixteen severe cases, four times in the thirty-one cases of medium severity and six times in the thirty-three light attacks.

It has so frequently been my experience to see severe pain in the ear or mastoid region, followed by a mild attack of facial paralysis, that I have been interested to record the location and character of the pain in most of the cases I have observed. In 187 cases, showing a complete or partial reaction of degeneration of the affected nerve, 56 gave a history of pain at the onset, while of 96 patients, who showed no change in the faradic irritability of the nerve, or a very slight decrease, and who recovered in from two to six weeks, 39 complained of aural or mastoid pain. On the whole, the pain experienced by those afflicted with the milder attacks has seemed quite as severe as that found in the severer cases, and in a number of instances has been sufficient to cause a series of sleepless nights.

The severity of the pain which may be experienced in facial paralysis, associated with herpes, is well illustrated by the following case. A woman, forty years of age, had suffered from a severe form of tic douloureux of the left face for several years. After having tried various forms of palliative treatment, an operation was decided upon. This was done by Dr. S. J. Mixter, who cut the second and third divisions of the trifacial nerve before they entered their respective foramina at the base of the skull, and plugged the foramina rotundum and ovale with amalgum, to prevent reunion. A good recovery was made from the operation and the patient spent a few weeks of comfort, when she began to have a return of paroxysms of pain. The pain now, however, was not in the malar and naso-labial region, but in the ear, and streaming down the jaw and into the neck. This pain increased and was so severe that the patient demanded another operation, being unable to sleep or eat on account of her suffering. Accordingly a second operation was performed, which showed that the third division of the fifth nerve actually had been divided, as the surgeon had claimed, and it was believed that the only means of obtaining a cure would be by division of the sensory root. This was not done, however, and three days later the patient was afflicted with a left-sided facial paralysis and a crop of herpetic clumps over the neck and left shoulder. It seems probable therefore that the pain the patient suffered before her second operation was due, not to trifacial neuralgia, but to an inflammatory process of her geniculate and cervical ganglia. She repeatedly described it, however, as being quite as severe as the old trifacial pain, though of a different quality.

In determining whether or not a continued course of treatment by galvanism, in cases of facial paralysis, presenting a reaction of degeneration, tends to the development of contracture, one is confronted with the difficulty of finding a standard, expressive of the degree of contracture, one wishes to describe. In classifying the results of treatment of such cases, I have made use of the term "marked," to indicate those cases with marked facial disfigurement as a result of the secondary contracture; "moderate," to indicate those in which the contracture is not absolutely disfiguring, but yet is apparent to the casual observation of an untrained eye; "slight," to denote those cases in which only a little accentuation of the lines of expression on the affected side can be detected.

In studying the relationship between treatment by galvanism and subsequent contracture, I have not included those cases presenting a partial degenerative reaction, since the degree of damage to nerve structure in these cases varies within wide limits. By making use of only the cases in which a complete R. D. was obtained, we are dealing with a degenerated nerve in all cases and can more fairly compare results.

In 49 cases of this class observed over two years after the onset of the paralysis, 28 had been through a four to eight months' treatment by galvanism, and of these 5 showed marked contracture, 6 moderate and 17 slight or none. Of the 21 patients who had not had galvanic treatment, 4 showed marked contracture, 10 moderate and 8 slight or none. From contrasting these two groups, it seems apparent that the administration of galvanism certainly plays no part in the causation of hemispasm secondary to facial paralysis. Whether or not the age of the patient has any bearing on the degree of contracture resulting is difficult to determine. Three of the patients included in the foregoing classification were under ten years of age and showed barely detectable contracture. On the other hand, there seems to be no more liability to severe hemispasm in those advanced in years than in the middle aged. Of course the degree of contraction has no direct bearing on the amount of motility regained for cases with only slight contracture, when the features are at rest, may show a marked impairment of motion.

Whether the secondary contracture of facial paralysis is due to a change in the muscle or in the nerve, is a point on which there is some variance of opinion. Hitzig believed the cause to be in the altered nerve cells in the nucleus and he has been supported in his view by Gowers (1). Purves Stewart (28) suggests that the real cause may be found in the incomplete regeneration of the affected muscle fibers. He bases his opinion on the work of Ioteyko (29), who holds that there exists in normal muscle two functionally distinct elements, viz., a fibrillar structure, which is irritable to faradism, and contracts with a quick twitch and sarcoplasm, which is less excitable and contracts slowly. This latter substance is in a constant state of semi-contraction, and maintains the normal tonicity of the muscle. Voluntary muscular movement is the result of a sort of a tetanus of fibrillary contractions, with an accentuation of the contraction of the sarcoplasm. When the muscle degenerates, the fibrillar substance is lost, hence the absence of faradic irritability of the muscle and the slow response to galvanism. If the regeneration is incomplete, the highly differentiated fibrillar substance is imperfectly reproduced and the sarcoplasm, being superabundant, keeps the muscle in a state of exaggerated tonus through the constant stimuli through the regenerated nerve. This explains why, when no regeneration of the nerve takes place, we have no appearance of contractures.

André Thomas, however, explains the development of secondary contracture as being a direct result of impulses, sent to the muscle through the irritation of the neuroma of the trunk of the nerve, which is formed in the process of irritation. He found in the pathological examination of a nerve taken from a patient who showed muscular contracture, as a result of previous facial paralysis, that above the geniculate ganglion was an area of variegated fibers. Some were hypertrophied and some very fine. At times several fine ones seemed within the same myeline sheath, and at times they appeared to bifurcate and give rise to many fibers. He suggests that the contracture may be due to pressure caused by this neuroma-like growth in the canal.

Such a condition as that described by André Thomas may not only explain the facial hemispasm following severe forms of facial paralysis, but it may also account for the associated movements, according to the theory of Lipschitz (30). He believes that in the process of regeneration the new axis cylinders may grow into different branches of the nerve, so that cells in the facial nucleus formerly innervating certain muscles may, through this change, come to supply all three branches of the nerve. According to the findings of André Thomas, it does not seem improbable that not only may an axis cylinder find a new destination, but that single axis cylinders may bifurcate to supply different muscles.

The temptation is strong, in going over such a series as we have been considering, to describe interesting phases of isolated cases, but my intention has been to adhere to the general questions considered here and to leave the details for another time.

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